BMC Ophthalmology

CASE REPORT Open Access

Pearl-type posterior capsule opacification as a possible predisposing factor for malignant glaucoma: a case report

Alexandre Barbé[1,2*], Gabin Momal[1], Alexis Cooren[1], Rafael Boucher[3] and Valentin Bachet[1]

Abstract

Background Malignant glaucoma (MG) is associated with a narrow angle or pseudoexfoliation (PEX)-induced laxity of the zonule. We describe a patient with massive posterior capsule opacification (PCO), i.e. Elschnig's Pearls (EP) and Soemmering's ring (SR) causing aqueous misdirection syndrome.

Case presentation A 78-year-old female was referred with rapidly progressive left ocular pain, redness, blurred vision of 20/100, and intraocular pressure (IOP) of 60 mmHg. She had undergone cataract surgery 5 years prior without complications, with a preoperative diagnosis of moderate capsular PEX syndrome. She was first treated by intravenous 250 mg acetazolamide along with maximal pressure-lowering drops, 1% pilocarpine and a patent laser iridotomy. Despite adding 500 cc of 10% mannitol, IOP remained high for 3 days. A shallow anterior chamber with angle closure, myopic shift and filling of the posterior chamber by massive PCO led us to conclude an aqueous misdirection syndrome. We promptly switched 1% pilocarpine to 1% atropine and performed a laser iridotomy enlargement with a posterior capsulotomy and anterior hyaloidotomy. This rapidly controlled the condition with posterior displacement of the intraocular lens (IOL) and fading of the high-IOP associated symptoms.

Conclusions Identifying malignant glaucoma could be challenging in the absence of immediate surgical circumstances. To our knowledge, this is the second description of PCO associated with MG. In this case, PCO was thought to narrow the iridocorneal angle and to cause a relative pupillary blockage, subsequently triggering aqueous humor misdirection to the vitreous and forward displacement of the iris-IOL diaphragm in the context of moderate PEX-induced zonular laxity.

Keywords Malignant glaucoma, Elschnig's pearls, Soemmering's ring, Posterior capsule opacification, Misdirection syndrome

*Correspondence:
Alexandre Barbé
alexandre.barbe.etu@univ-lille.fr
[1]Service d'ophtalmologie, Centre hospitalier Universitaire de Lille, Hôpital Huriez, Lille, France
[2]Université de Lille, faculté de médecine Henri Warembourg, Lille, France
[3]Service d'ophtalmologie, Centre hospitalier du Kremlin-Bicêtre, APHP, Paris, France

Introduction

Malignant glaucoma was first described by von Graefe in 1869 as an aggressive form of postoperative glaucoma resistant to medical treatment and resulting in blindness [1]. It mostly occurs after filtration surgery for angle-closure glaucoma, but has also been reported after phacoemulsification, laser iridotomy, laser capsulotomy, cyclophotocoagulation, trabeculectomy, scleral flap suture lysis, trabeculectomy bleb needling [2–4]. Moreover, it can be induced by miotics or occurs spontaneously [3], [4]. MG is classified among the angle-closure glaucoma. The diagnosis is based on high IOP associated with shallow central and peripheral anterior chamber, myopic shift [2] in the absence of acute pupillary block (patent iridotomy) and exclusion of posterior segment pathologies (suprachoroidal hemorrhage and choroidal effusions) [5–7]. The pathogenesis remains unclear. It is believed to be the consequence of an anterior rotation of the ciliary body leading to misdirection of aqueous humor flow into the vitreous cavity, resulting in forward displacement of the iris-lens diaphragm in an anatomically predisposed eye [6–10]. In this report, we describe a possible association between pearl-type PCO (EP and SR) and angle closure leading to malignant glaucoma in a pseudophakic patient.

Results

A 78-year-old woman went through bilateral phacoemulsification in 2015. Preoperative findings were bilateral PEX syndrome (Fig. 1A). No complications occurred during the operation, nor was the post-operative IOP elevated (14–19 mmHg from 2016 to 2019, central corneal thickness = 510 µm in both eyes -BE). She presented to the emergency unit in 2020 with crucial pain, nausea and vomiting, redness and blurred vision in the left eye (LE) ongoing for 3 days. Noticeable was a myopic shift LE: -0.25 spherical diopters (D) in March 2019 and– 4.00 D on the day of the crisis whereas no change was noted in the right eye (RE). The best-corrected visual acuity (BCVA) was 20/20 RE and 20/100 LE. IOP was 18 mmHg RE and 60 mmHg LE. The initial slit lamp examination (SLE) of the left eye revealed a shallow anterior chamber (van herick grade 1, Fig. 1B), corneal edema, pericorneal vascular injection, no sign of anterior chamber inflammation and PCIOL in place with EP (Fig. 1C, D red dotted circle). The pupil was unreactive to light, mid-dilated. The iridocorneal angle was 360°-closed and the fundus was blurry but seemed unremarkable. RE examination was unremarkable except for the presence of EP.

B-mode echography ruled out choroidal effusion or suprachoroidal hemorrhage. Ultrasound biomicroscopy (Fig. 2A, B) and anterior segment optical coherence tomography (OCT) (Fig. 2C, D) confirmed the angle closure LE and the presence of Elschnig's pearls (Fig. 2A, *red dotted arrow*) and Soemmering's ring circularly attached to the iris (Fig. 2B, *blue arrow*). No "iris bombé" sign nor choroidal effusion were found. Of note a mass effect was detected on the iris curve (Fig. 2A, *red dotted arrow*).

Our first hypothesis as the main angle-closure mechanism was pupillary-block and the following treatment was initiated: 250 mg intravenous acetazolamide (SANOFI-AVENTIS FRANCE) with prompt addition of 1% iopidine (apraclonidine, ESSENTIAL PHARMA LIMITED), geltim LP (timolol maleate 1 mg/g, THEA), lumigan (bimatoprost 0.3 mg/mL, ALLERGAN FRANCE) and trusopt (dorzolamide 20 mg/mL, SANTEN SAS) every 5 min for 6 h without success. Drops of 1% pilocarpine (pilocarpine 1% FAURE, EUROPHTA) were added and a Nd: YAG laser iridotomy (Abraham iridectomy Lens, 8-10 mJ) was performed with no significant result. 1% pilocarpine drops were thus rapidly switched for 1% atropine (atropine 1% FAURE, EUROPHTA) drops and intravenous 500 cc of 10% mannitol (mannitol, AGUETTANT) were added. Despite maximal medical

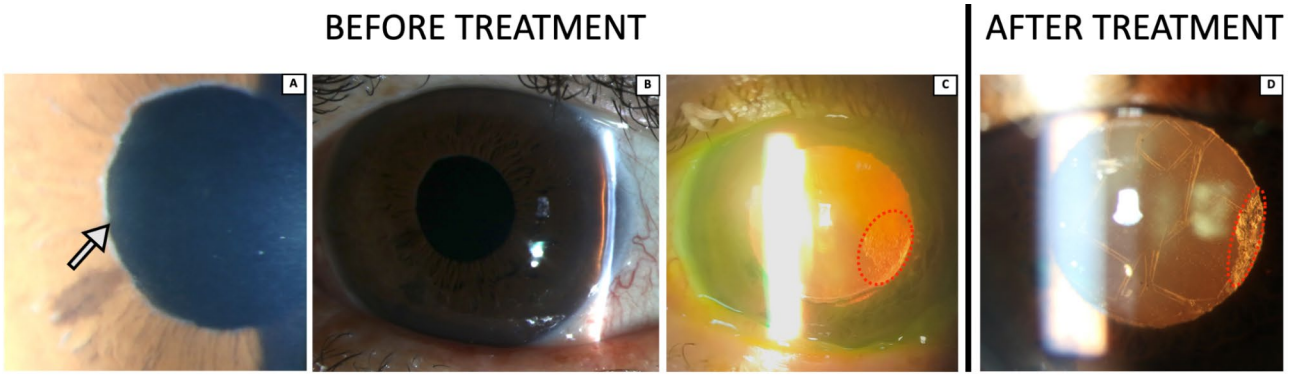

Fig. 1 Slit-lamp biomicroscopy images of the patient with malignant glaucoma (MG). **A**: pseudoexfloliation materials deposits on iris pupil (white arrow), known as a risk factor of zonular laxity and of the iris-lens forward displacement during malignant glaucoma. **B**: before treatment, shallow anterior chamber (van herick grade 1). **C**: posterior chamber intraocular lens with Elschnig's pearls (red dotted circle). **D**: Nd:YAG capsulotomy / anterior hyaloidotomy allowing aqueous humor to circulate to the deepened anterior chamber. Note the Elschnig's pearls (red dotted circle)

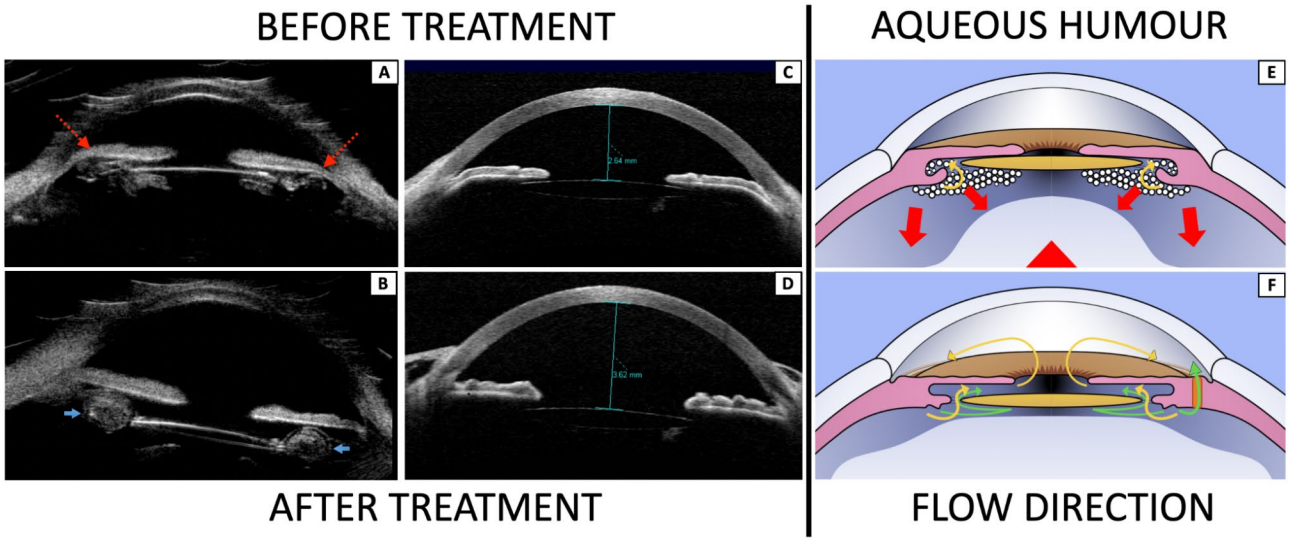

Fig. 2 Anterior segment images of the left eye before (**A**, **C**) and after treatment (**B**, **D**) of the malignant glaucoma (left panel) and hypothetical pathophysiology of the phenomenon (right panel: **E**, **F**). **A** and **B**: ultrasound biomicroscopy (UBM) showing surrounding mass, i.e. Elschnig's pearls (red dotted arrow) and Soemmering's ring (blue arrow) aside the posterior chamber implant. No choroidal effusion or complete pupillary block (« iris bombé » sign). Note that there is a mass effect of the PCO seen on the iris curve pointed by the red dotted arrow (**A**). Deepening of the anterior chamber (from 2.6 mm to 3.6 mm anterior chamber depth) and posterior displacement of the ciliary body and PCO after treatment (**B**). **C** and **D**: anterior segment OCT with deepening of the anterior chamber and opening of the irido-corneal angle after treatment (**D**). **E**: hypothesis of misdirection of aqueous humor (AH) in case of EP with "plateau-like" and relative blockage of AH in the posterior chamber, AH accumulating posteriorly increasing vitreous pressure thus displacing forward the iris-IOL diaphragm. This vicious circle leads to the closure of the iridocorneal angle. **F**: physiological aqueous humor circulation (yellow arrow) in a pseudophakic patient and alternative route after Nd: YAG capsulotomy / anterior hyaloidotomy and irido-zonulo-hyaloidotomy (green arrows, i.e. in our case after treatment)

therapy, IOP remained elevated at 60 mmHg LE on the third day. Eventually, the patient underwent iridotomy enlargement and Nd: YAG laser capsulotomy with disruption of the anterior hyaloid membrane (circular pattern to open wide and avoid retinal tears, Lens Abraham capsulotomy, 5-6mJ, *Fig. 1C*). The pain disappeared, IOP dropped to 19 mmHg LE and SLE normalized: fading of corneal edema and pericorneal vascular injection, anterior chamber deepening and opening of the iridocorneal angle (Fig. 2A-D*).* The gonioscopy showed a fully open angle without synechia. Refraction was – 1,25 D with a visual acuity of 20/25 two weeks after, close to her BCVA before. No subsequent damage was noted on peripapillary OCT at one month.

Discussion

Malignant glaucoma, also known as misdirection of aqueous humor or ciliary block, is a rare condition. It is characterized by elevated IOP, shallow anterior chamber, myopic shift [2], absence of suprachoroidal effusion [11], no response to miotics or peripheral iridectomy, and good response to cycloplegics [2, 5–7, 12]. It is associated with anatomical conditions (axial hyperopia, a narrow angle [9, 13], ciliary sulcus or plateau iris configuration [14]) or PEX-induced laxity of the zonule in the sunset syndrome [13] and in anterior IOL or capsular tension ring displacement [2, 13–15]. MG is considered a deviation of aqueous flow in the vitreous body increasing posterior pressure and leading to a forward displacement of the iris-lens diaphragm [8, 16]. It mostly occurs after filtration surgery in hyperopic eyes but can follow laser iridotomy/capsulotomy, cyclophotocoagulation, cataract surgery [2], trabeculectomy scleral flap suture lysis, and trabeculectomy bleb needling [3, 4]. It appears with various latencies, from the immediate postoperative period up to 16 years after surgery [2]. Medical treatment consists of cycloplegic agents (relaxing the ciliary muscle) and aqueous suppressants (reducing the vitreous pressure) [17]. In refractory cases, laser or surgery may be of great help. A patent laser iridotomy may shrink the ciliary processes. Prophylactic iridotomy/iridectomy in the fellow eye should be considered if the irido-corneal angle is narrow before any surgical intervention [17]. Pars plana vitrectomy or combined vitrectomy-iridectomy-zonulectomy (and phacoemulsification if needed) have also been proposed [2, 17].

Our patient had normal axial length, no filtration surgery, no subluxation of PCIOL and a fully open iridocorneal angle in 2018. Her risk factors for MG were phacoemulsification 5 years prior and moderate PEX syndrome. An acute complete pupillary block was ruled out as no "iris bombé" was present on ultrasound biomicroscopy images and neither pilocarpine nor patent Nd: YAG iridotomy decreased the IOP. Only atropine,

laser iridotomy enlargement and posterior capsulotomy with anterior hyaloidotomy were rapidly successful. In this case, the first attempt of laser iridotomy was not efficient as the pressure came from the vitreous cavity and was blocked by pearl-type PCO with mass effect on the iris curve causing a relative pupillary block (Fig. 2A, *red dotted arrow*). Laser iridotomy enlargement acted as an irido-zonulo-hyaloidotomy, allowing the aqueous flow between the posterior segment and anterior chamber (Fig. 2F, *green arrow in the orange channel*). Nd: YAG laser also helped this release by rupturing the posterior capsule and anterior hyaloid membrane connecting the vitreous body with the posterior chamber and thus the anterior chamber (Fig. 2F, *green arrows*). Myopic shift and ultrasound biomicroscopy images showing a filling of the posterior chamber by massive PCO ("plateau-like" configuration of the iris) and anterior displacement of the iris-IOL diaphragm led us to the track of MG. Indeed, PCO was responsible for this "plateau-like" configuration [18, 19] and was thought to trigger a relative pupillary block [15, 20–22] with aqueous flow misdirection to the posterior segment leading to increased vitreous pressure, thus generating anterior displacement of the iris-lens diaphragm. This was most likely facilitated by a predisposed PEX-induced zonular weakness. This chronic aqueous humor misdirection by PCO might be another predisposing factor for malignant glaucoma. It should be taken into consideration with an anticipated Nd: YAG laser capsulotomy before planning any surgery.

Posterior capsule opacification is developed from residual lens epithelial cells of the cataract [23]. It is clinically divided into two categories: the fibrosis type (proliferation, migration, epithelial to mesenchymal transition of the lens epithelial cells and collagen deposition) and the pearl-type (regeneration of crystallin-expressing lenticular fibers). Lens epithelial cells migrate and proliferate between the posterior capsule and the IOL, first forming syncytial posterior capsular opacification and later Elschnig's pearls or Soemmering's ring [23, 24]. There has been a single report of MG in the context of pearl-type PCO in a patient with nanophthalmic eyes undergoing phacoemulsification and trabeculectomy complicated by immediate MG [25]. Treatment consisted of an anterior vitrectomy and peripheral lens capsular hole with peripheral iridectomy [25]. Two months later, Elschnig's pearls caused closure of the capsular hole with recurrence of MG treated by enlargement of the lens capsular hole. In this extremely predisposed patient for MG, Elschnig's pearls acted as a direct obstacle to aqueous humor flow. In our case, pearl-type PCO was also thought to trigger MG by mechanical obstruction but in different ways: reducing the physiological flow of the aqueous humor behind the iris (relative pupillary blockage) and narrowing the iridocorneal angle (with a "plateau-like" configuration). To our knowledge, this is the second report of pearl-type PCO associated with a malignant glaucoma case.

Abbreviations

BCVA	Best corrected visual acuity
BE	Both eyes
D	Diopters
EP	Elschnig's pearls
IOP	Intraocular pressure
IOL	Intraocular lens
LE	Left eye
MG	Malignant glaucoma
OCT	Optical coherence tomography
PCIOL	Posterior chamber intraocular lens
PCO	Posterior capsule opacification
PEX	Pseudo-exfoliation syndrome
RE	Right eye
SLE	Slit lamp examination
SR	Soemmering's ring

Acknowledgements

We would like to thank Clément Barbé https://clement-barbe.com/links for the illustration (graphic designer of the Fig. 2E, F).

Author contributions

AB collected, analyzed, and interpreted the clinical data. AB and GM wrote the first draft of this manuscript. AB, GM, RB, AC and VB contributed to the supervision of the manuscript and medical care of the patient. All authors have reviewed and approved the final version of the manuscript.

Funding

No funding or grant support.

Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent was obtained from the relative (daughter) of the patient for the publication of this report and any accompanying images.

Competing interests

The authors declare no competing interests.

Received: 3 June 2024 / Accepted: 6 January 2025
Published online: 28 March 2025

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